CASE REPORTS

- **Pulmonary Atresia with Patent Interauricular and Interventricular Septal Defects Causing Functional Cor Biloculare in a Patient with Dissociated Dextrocardia**
- **Hexamethonium Contributing to Fatal Shock in Hypertensive Epistaxis**

Pulmonary Atresia with Patent Interauricular And Interventricular Septal Defects Causing Functional Cor Biloculare in a Patient with Dissociated Dextrocardia

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CONGENITAL LESIONS OF THE HEART are by no means rare. Such defects were noted in 1,171 of 40,130 cases in which autopsy was done at the Los Angeles County General Hospital between 1918 and 1948, an incidence of 2.9 per cent. The commonest malformation was patent foramen ovale, or interauricular septal defect, which was observed in 255 cases (0.5 per cent). Next were patent ductus arteriosus, noted in 135 cases (0.2 per cent), and interventricular septal defect in 126 cases (0.2 per cent). Congenital dextrocardia was observed in only four cases, and in three of them it was associated with general situs inversus.

In acquired dextrocardia there is no transposition of other organs; but in congenital dextrocardia the heart usually is dextroverted as though in mirror image and usually in the rest of the body also there is situs inversus—the stomach and spleen on the right side and the liver and appendix on the left. Rarely are other congenital defects of the heart associated with congenital dextrocardia when it is a part of generalized situs inversus. However, Kartagener's triad—sinusitis, bronchiectasis and dextrocardia—has been frequently reported. In the much rarer congenital dextrocardia without situs inversus of other organs, however, multiple and extensive congenital malformations of the heart are not uncommon. In the following case of congenital dextraposition of the heart there was extensive structural malformation.

REPORT OF A CASE

A 12-year-old Mexican girl was admitted in emergency to Olive View Sanatorium in November 1949 because of hemoptysis following what was described as severe upper respiratory tract infection with frequent cough and pain in the chest. In x-ray films taken upon admittance the patient was observed to have far advanced bilateral pulmonary tuberculosis with a thin-walled cavity 2 cm. in diameter in the right mid-lung field. The sputum contained acid-fast bacilli.

A "blue baby" at birth, the patient had had increasing cyanosis and shortness of breath after only slight physical exertion. She was physically retarded (had not walked until four years of age) but mentally alert and seemed to have the intelligence normal for children of her age. She had had pertussis when two years of age and measles at ten years. There was no history of rheumatic stigmata or prolonged febrile disease. The patient had been observed periodically in the cardiac clinic of another hospital from January 1939 to July 1947 with diagnosis of congenital cardiac abnormality of cyanotic type, and there were no episodes of cardiac failure or thromboembolic phenomenon during that time. When three years of age she had repeatedly visited a patient with active tuberculosis, and it is believed that she had a previous primary tuberculous infection which caused enlarged mediastinal lymph nodes and an atelectatic left upper lobe.

Upon physical examination the patient was observed to be underdeveloped, poorly nourished and cyanotic. The body weight was 47 pounds. There was pronounced clubbing of the fingers and toes. The temperature was 102° F., the pulse rate 126, the blood pressure in the arms 110 mm. of mercury systolic and 70 mm. diastolic, and in the legs 130 mm. and 80 mm. respectively. The skin and mucous membranes of the head and neck and extremities were deep purple. There was slight distention of the veins in the neck. Symmetrical underdevelopment of the chest was noted and there was a slight bulging of the lower anterior portion of the chest. Except for an area of dullness posteriorly over the left upper lobe of the lungs, response to percussion over the lung fields was essentially normal. No rales were heard upon auscultation. Upon examination of the heart the point of maximum intensity was located in the fourth interspace to the right of the sternum and the area of retrocardiac dullness extended 4 cm. to the right and 2 cm. to the left of the sternum. There was a loud, rasping, grade IV systolic murmur heard best in the second interspace to the left of the

sternum, and it was accompanied by a systolic thrill. The murmur radiated down the left side of the sternum and upward toward the left clavicle, but was only faintly heard in the neck, axilla and back. P^2 was snapping. The liver, palpable three fingers' breadth below the left costal margin, was nontender. No masses or areas of tenderness were noted in palpation of the abdomen.

Erythrocytes numbered 7,190,000 per cu. mm. of blood and the hemoglobin value was 122 per cent. The sedimentation rate was only moderately accelerated. The patient had vital capacity of 900 cc.

Upon fluoroscopic examination the cardiac silhouette was observed to be predominantly in the right side of the chest, and active pulsations which seemed to come from an enlarged right ventricle were noted. Electrocardiographic tracings were consistent with dextroposition of the heart with right ventricular preponderance. Angiocardiographic observations and results of cardiac catheterization done at the Los Angeles County General Hospital in December 1950 were consistent with truncus arteriosus (see Figures 1, 2, 3 and 4). Exploratory operation

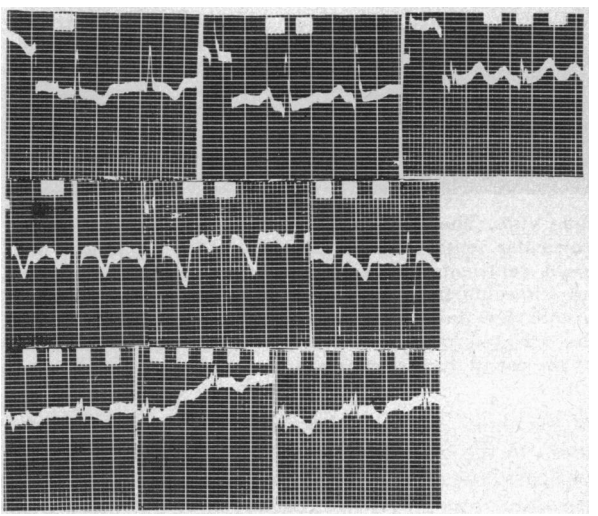

Figure 1.—Electrocardiographic abnormalities consistent with right heart preponderance and strain.

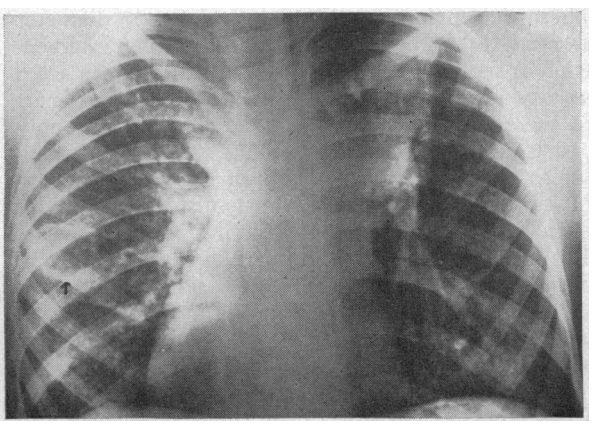

Figure 2.—X-ray of chest indicating dextroposition of the heart, thin-walled tuberculous cavity in right lung, and soft follicular type spread in both lung fields. The left upper lobe also has a fibrocalcific lesion with partial atelectasis.

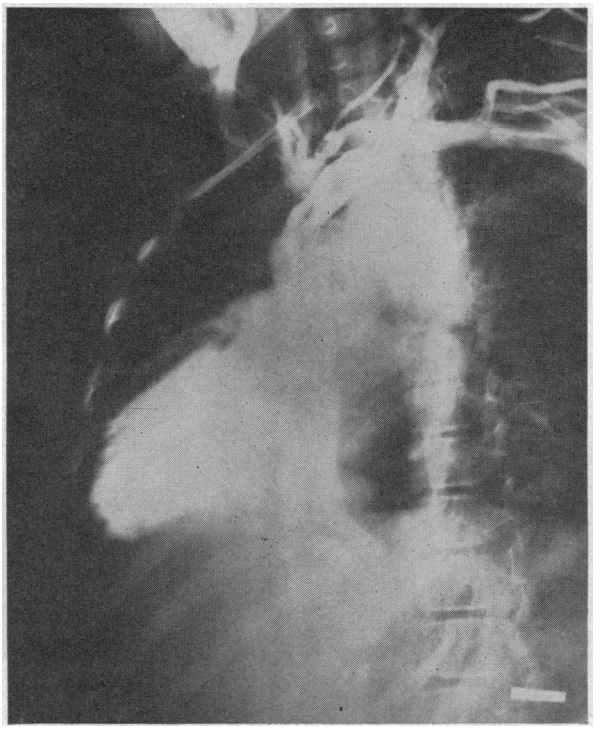

Figure 3.—Angiocardiogram. Dye filling the right auricle and ventricle and remnants of the superior and inferior vena cava.

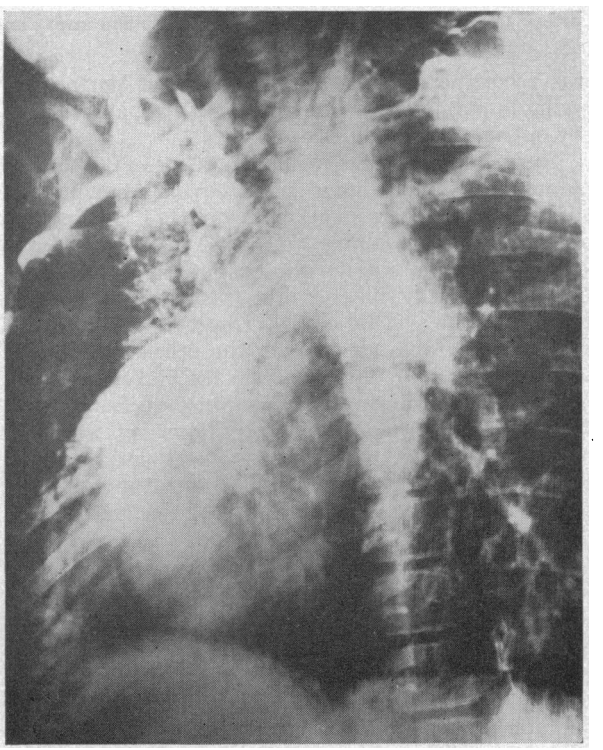

Figure 4.—Angiocardiogram. Residual dye in the right heart. The aorta can now be faintly visualized.

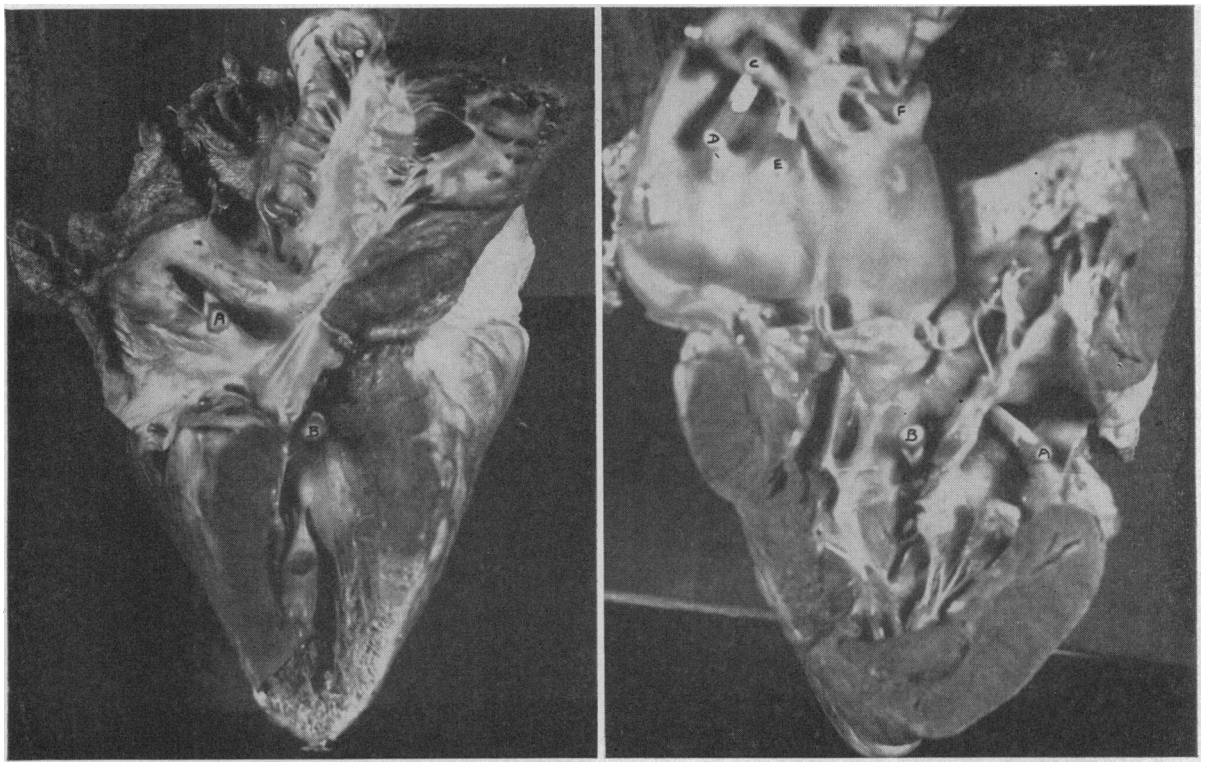

Figure 5—*Left*—Pathological specimen of heart, right oblique view. The right auricle and ventricle are reflected to show the openings of (A) the interauricular and (B) interventricular septal defects. The tricuspid valve is split in order to better visualize these openings. The thickness of the right ventricular musculature approximates that of the left ventricle. There was no pulmonary aorta and only a few shreds could be found at the site where the pulmonary aorta usually leaves the right ventricle. *Right*—In the left oblique view the septal defects are again visualized (A) interauricular and (B) interventricular. The systemic aorta is here reflected in order to demonstrate (C) the left carotid artery (D) right in nominate (F) left subclavian artery and (E) the patent ductus arteriosus.

was recommended at this time but was postponed owing to pulmonary hemorrhage with apparent miliary spread of tuberculosis.

Thereafter there was further increase in the tuberculous area in the lungs and frequent hemoptysis. Cyanosis was persistent but variable, becoming worse during coughing or crying spells. In July 1950, following miliary dissemination, the patient became critically ill and dihydrostreptomycin was given. There was good clinical response and in x-ray films considerable clearing in the areas of dissemination was noted. The cavity in the right mid-lung became larger, however, and because of the risk of further hemorrhages and pulmonary spread, pneumothorax on the right side was carried out. Some months later, two days after pneumothorax refill, massive pulmonary hemorrhage occurred and the patient died.

Postmorten examination. The body, about 50 inches long, weighed about 50 pounds. The skin and sclera were bluish, but there was only slight dependent lividity. The fingernails were curved and cyanotic. There was no edema.

The right pleural cavity, which contained about one liter of air, was traversed by numerous "string" adhesions. Diffuse fibrous adhesions that were difficult to separate held the left lung firmly to the wall of the chest. The heart lay in the midline, extending more to the right than to the left. No lesions of the pericardium, epicardium, myocardium or endocardium were noted macroscopically, but there was extreme abnormality in the structure of the entire heart (Figure 5). The left ventricular muscle was as thick as the right; each was about 8 mm. thick. There were patent openings about 1 cm. in diameter through both the interauricular and the interventricular septa. Functionally the heart must have acted as a two-chamber heart. No opening from the heart to the pulmonary artery could be found, the only exit from the right ventricle being through the opening in the interventricular septum. The superior and inferior vena cava opened normally into the right auricle and the pulmonary veins into the left auricle. The aorta arose from the left ventricle in the normal position. Along its base arose several occluded shreds of an atretic vessel running to the pulmonary artery, but the supply of blood to the pulmonary artery came entirely from a large patent ductus arising in the ascending aorta just below the arch about 1 cm. from the left subclavian artery. The aorta and its major branches were large and thick, and there were a few atheromatous plaques near the aortic valve, but no other gross abnormalities were observed. The heart weighed 175 grams.

Both lungs were crepitant for the most part, but there was an area of consolidation around a cavity about 2 centimeters in diameter in the lower part of the upper right lobe. The cavity was thin-walled and completely filled with one clot of blood. There were also scattered clots in other parts of the lung, but no tuberculous lesions were seen except for the cavity on the right side. The right lung weighed 200 gm., the left 225 gm. The upper part of the left upper lobe showed atelectatic consolidation with fibrotic replacement of the parenchyma and dilation of the bronchi with multiple saccular cavities measuring 0.5 to 2 cm. in diameter. The cavities were completely filled with fresh clotted blood. The source of hemorrhage was not found, although the cavity and bronchial walls appeared hyperemic. There were scattered clots of blood in other parts of the left lung and some fresher tuberculous infiltration in the upper part of the left upper lobe. The liver, spleen, kidneys and alimentary tract were in normal position and no congenital abnormalities were observed in them.

The probable cause of death was hemorrhage from a bronchiectatic cavity in the left lung, possibly precipitated by increased intrapulmonary pressure secondary to pneumothorax and pulmonary hypertension owing to congenital malformation of the heart.

DISCUSSION

Reports of several somewhat similar cases[2, 3, 4] have been published. In each case there was obvious clinical evidence of congenital cardiac defect—cyanosis from birth and clubbing of fingers. In those cases, as in the one herein reported, enlargement of the heart to the right and palpable thrill were noted upon physical examination, and in most of them x-ray films showed right sided position of the heart and transverse enlargement. Electrocardiographic tracings showed right axis deviation and other abnormalities. Cardiac catheterization[1, 5] and study of chemical factors in the blood were carried out only in one previous case. In that case x-ray films taken in rapid series after intravenous injection of Diodrast helped to reveal the true nature of the cardiac defect.

SUMMARY

In a case of pulmonary atresia with interauricular and interventricular septal defects with dissociated dextrocardia, clinical, electrocardiographic, roentgenographic and cardiac catheterization findings were correlated with the conditions observed at autopsy.

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Hexamethonium Contributing to Fatal Shock in Hypertensive Epistaxis

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HEXAMETHONIUM, a powerful ganglionic blocking agent, is one of the new drugs that have appeared within the last few years for management of hypertension. According to Hilker and associates,[2] hexamethonium action is unpredictable and it causes wide variations in blood pressure when administered by parenteral route. Even oral administration is not completely safe. Toxic manifestations of hexamethonium are numerous—orthostatic hypotension, paralytic ileus, paralysis of the bladder, circulatory collapse, various gastrointestinal upsets, headache, visual disturbances, dyspnea and fatigue, among others.

Grimson and co-workers[1] treated 103 hypertensive patients with hexamethonium and observed three deaths from cardiovascular accident and uremia. These investigators emphasized both sympatholytic and parasympatholytic effects of this drug. Wilkins[5] detected ischemic changes in an electrocardiogram following parenteral administration of 10 mg. of hexamethonium. Morrison[3] described a case in which the patient died of coronary thrombosis and apparently death was precipitated by hexamethonium. Rytand[4] said that hexamethonium is suitable for only a few patients, and that it fails in persons with renal insufficiency, who most need help.

The present report is of a patient with severe essential hypertension and intractable epistaxis who was treated by hexamethonium bromide parenterally in a last desperate measure to control the blood pressure. Overwhelming circulatory collapse occurred and eventually the patient died.

REPORT OF A CASE

A 61-year-old white man was admitted to hospital June 15, 1952, with a history of high blood pressure for many years. He fainted on the day of entry and was feeling weak and dizzy. On examination, the blood pressure was 230/140 mm. of mercury, the